## IN THE CLAIMS:

This listing of claims replaces all prior versions and listings of claims in the application:

## Listing of Claims

- 1-25. (Canceled)
- 26. (Previously presented) A transgenic D. melanogaster comprising
- a transgene containing a plurality of CAG's and at least one CAA sequence encoding a polyglutamine repeat sequence operably linked to a constitutive, regulatable, or tissue specific expression control element, wherein the tissue specific expression control element is selected from the group consisting of Appl, rhodopsin 1 promoter, and GLASS transcription factor element, and wherein the transgene produces polyglutamine toxicity in the transgenic *D. melanogaster*.
- 27-28 (Canceled)
- 29. (Previously Presented) The *D. melanogaster* of claim 26, wherein the number of CAG's to CAA's is in ratio of between about 1:1 and 2:1.
- 30. (Previously Presented) The *D. melanogaster* of claim 26, wherein the number of CAG's to CAA's is in ratio of between about 2:1 and 5:1.

- 31. (Previously Presented) The *D. melanogaster* of claim 26, wherein the number of CAG's to CAA's is in ratio of between about 5:1 and 10:1.
- 32. (Previously Presented) The *D. melanogaster* of claim 26, wherein the number of CAG's to CAA's is in ratio of between about 10:1 and 50:1.
- 33. (Canceled)
- 34. (Previously Presented) The *D. melanogaster* of claim 26, wherein the tissue specific expression control element confers neural, retinal, muscle or mesoderm cell expression.
- 35-36. (Canceled)
- 37. (Previously Presented) The *D. melanogaster* of claim 26, wherein the polyglutamine sequence is between about 50 and 100 amino acids in length.
- 38. (Previously Presented) The *D. melanogaster* of claim 26, wherein the polyglutamine sequence is between about 100 and 200 amino acids in length.
- 39. (Previously Presented) The *D. melanogaster* of claim 26, wherein the polyglutamine sequence is between about 50 and 200 amino acids in length.

40. (Previously Presented) The *D. melanogaster* of claim 26, wherein the polyglutamine sequence further comprises a tag.

## 41. (Canceled)

42. (Currently Amended) The D. melanogaster of claim 26, wherein the Drosophila further comprises a marker sequence inserted into its genomic DNA, wherein the marker is located adjacent to a gene selected from the group consisting of a HDJ1 gene, a TPR2 gene, and a MLF gene a gene or inserted into a gene whose expression or activity increases or decreases polyglutamine toxicity in the animal, and wherein the marker sequence comprises an inducible upstream activating sequence, a minimal promoter sequence and 5' and 3' P transposon elements containing terminal inverted repeats.

## 43. (Canceled)

- 44. (Currently Amended) The D. melanogaster of claim  $\underline{42}$   $\underline{43}$ , wherein the gene is HDJ1.
- 45. (Currently Amended) The D. melanogaster of claim  $\underline{42}$   $\underline{43}$ , wherein the gene is TPR2.
- 46. (Currently Amended) The D. melanogaster of claim  $\underline{42}$   $\underline{43}$ , wherein the marker sequence is near an MLF gene.
- 47. (Withdrawn) A method for identifying a compound that modulates polyglutamine toxicity in an animal comprising:

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- (a) contacting the animal of claim 41 with a test compound; and
- (b) determining whether the test compound increases or decreases polyglutamine toxicity in the animal, where increased or decreased polyglutamine toxicity identifies the test compound as a compound that modulates polyglutamine toxicity.
- 48. (Withdrawn) The method of claim 47, wherein the compound is present in the animal's food or drink.
- 49. (Withdrawn) The method of claim 47, wherein the compound is administered to a tissue or organ of the animal.
- 50. (Previously Presented) A method of producing a transgenic D. melanogaster characterized by suppressed polyglutamine toxicity comprising:
- (a) transforming a *D. melanogaster* embryo or fertilized egg with a transgene comprising a plurality of CAA and CAG sequences encoding a polyglutamine sequence operably linked to a constitutive, regulatable, or tissue specific expression control element, wherein the tissue specific expression control element is selected from the group consisting of Appl, rhodopsin 1 promoter, and GLASS transcription factor element and
- (b) selecting a *D. melanogaster* that exhibits polyglutamine toxicity.
- 51. (Withdrawn) An isolated polynucleotide sequence having about 65% or more identity to a Drosophila TPR2 (dTPR2) sequence set forth as SEQ. ID NO:2 and which encodes a polypeptide that decreases polyglutamine toxicity, with the proviso that the

sequence is distinct from the EST sequences set forth in Figure 11.

- 52. (Withdrawn) The polynucleotide sequence of claim 51, wherein the sequence encodes a subsequence of TPR2 that decreases polyglutamine toxicity.
- 53. (Withdrawn) The polynucleotide sequence of claim 51 operatively linked to an expression control element.
- 54. (Withdrawn) An isolated polynucleotide sequence that hybridizes under stringent conditions to a Drosophila TPR2 (dTPR2) sequence set forth as SEQ. ID NO:2, with the proviso that the sequence is distinct from the EST sequences set forth in Figure 11.
- 55. (Withdrawn) The polynucleotide sequence of claim 54, wherein the sequence comprises a polynucleotide having 20 or more contiguous nucleotides.
- 56. (Withdrawn) The polynucleotide sequence of claim 54, wherein the sequence comprises a polynucleotide having 50 or more contiguous nucleotides.
- 57. (Withdrawn) An isolated polynucleotide sequence having about 65% or more identity to a Drosophila MLF (dMLF) sequence set forth as SEQ. ID NO:4 and which encodes a polypeptide that decreases polyglutamine toxicity, with the proviso that the sequence is distinct from the EST sequences set forth in Figure 12.

- 58. (Withdrawn) The polynucleotide sequence of claim 57, wherein the sequence encodes a subsequence of MLF that decreases polyglutamine toxicity.
- 59. (Withdrawn) The polynucleotide sequence of claim 57 operatively linked to an expression control element.
- 60. (Withdrawn) An isolated polynucleotide sequence that hybridizes under stringent conditions to a Drosophila MLF (dMLF) sequence set forth as SEQ. ID NO:4, with the proviso that the sequence is distinct from the EST sequences set forth in Figure 12.
- 61. (Withdrawn) The polynucleotide sequence of claim 60, wherein the sequence comprises a polynucleotide having 20 or more contiguous nucleotides.
- 62. (Withdrawn) The polynucleotide sequence of claim 60, wherein the sequence comprises a polynucleotide having 50 or more contiguous nucleotides.
- 63. (Withdrawn) A composition comprising a polynucleotide sequence encoding a human MLF polypeptide operatively linked to an expression control element in a pharmaceutically acceptable carrier.
- 64. (Withdrawn) A composition comprising a polynucleotide sequence encoding a human TPR2 polypeptide operatively linked to

an expression control element in a pharmaceutically acceptable carrier.

- 65. (Withdrawn) A method of increasing survival of a cell having polyglutamine toxicity, comprising contacting the cell with an amount of TPR2 or MLF polypeptide sequence or a polynucleotide sequence TPR2 or MLF polypeptide to increase survival of the cell.
- 66. (Withdrawn) A method of decreasing apoptosis of a cell, comprising contacting the cell with an amount of TPR2 or MLF polypeptide sequence or a polynucleotide sequence TPR2 or MLF polypeptide to decrease apoptosis of the cell.
- 67. (Withdrawn) A method of decreasing polyglutamine toxicity in a cell having or susceptible to polyglutamine toxicity, comprising contacting the cell with an amount of J domain containing polypeptide, TPR2 or MLF polypeptide sequence, or a polynucleotide sequence encoding the J domain containing polypeptide, TPR2 or MLF polypeptide sequence to decrease polyglutamine toxicity in the cell.
- 68. (Withdrawn) The method of claim 67, wherein the cell is a neural, retinal, muscle or mesoderm cell.
- 69. (Withdrawn) The method of claim 67, wherein the toxicity is decreased by decreasing cell death or increasing cell survival.

- 70. (Withdrawn) A method of decreasing polyglutamine toxicity in a tissue or organ of a subject having or at risk polyglutamine toxicity, comprising contacting the tissue or organ with an amount of a J domain containing polypeptide, a TPR2 or MLF polypeptide sequence, or a polynucleotide sequence encoding the J domain containing polypeptide, TPR2 or MLF polypeptide, to decrease polyglutamine toxicity in the tissue or organ of the subject.
- 71. (Withdrawn) The method of claim 70, wherein the tissue is brain, eye, muscle or mesoderm.
- 72. (Withdrawn) A method of decreasing the severity of a frontotemporal dementia, prion disease, polyglutamine disorder or protein aggregation disorder in a subject having or at risk of a frontotemporal dementia, prion disease, polyglutamine disorder or protein aggregation disorder, comprising administering to the subject an amount of J domain containing polypeptide, a TPR2 or MLF polypeptide sequence, or a polynucleotide sequence encoding the J domain containing polypeptide, TPR2 or MLF polypeptide, to decease the severity of the frontotemporal dementia, prion disease, polyglutamine disorder or protein aggregation disorder in the subject.
- 73. (Withdrawn) The method of claim 72, wherein the method comprises prophylactic administration.
- 74. (Withdrawn) The method of claim 72, wherein the disorder is a neurological or muscle disorder.

- 75. (Withdrawn) The method of claim 72, wherein the disorder impairs long term or short term memory or coordination of the subject.
- 76. (Withdrawn) The method of claim 72, wherein the disorder is characterized by the presence of protein aggregates, amyloid plaques, degeneration or atrophy in an affected tissue or organ.
- 77. (Withdrawn) The method of claim 72, wherein the disorder is selected from the group consisting of Alzheimer's disease, Parkinson's disease, Creutzfeldt-Jacob's disease (CJD), bovine spongiform encephalopathy, Huntington's disease (HD), Machado-Joseph disease (MJD), Spinocerebellar ataxias (SCA), dentatorubropallidoluysian atophy (DRPLA), Kennedy's disease, stroke and head trauma.
- 78. (Withdrawn) The method of claim 72, wherein the severity is decreased by decreasing cell death or increasing cell survival.
- 79. (Withdrawn) The method of claim 72, wherein the severity is decreased by decreasing protein aggregation.